Rheumatic Fever

Section 1: ABOUT THE DISEASE

Note: This chapter focuses on rheumatic fever, a complication of infection with Streptococcus pyogenes (group A streptococci [GAS]). For information about invasive GAS infections, refer to the chapter titled "Group A Streptococcus (Invasive)."

A. Etiologic Agent

Rheumatic fever is a delayed manifestation of bacterial infection with group A streptococci (GAS), also known as *Streptococcus pyogenes*. There are over 130 serotypes of GAS. Epidemiologic evidence suggests an association between certain serotypes (those which tend to cause upper respiratory infections) and rheumatic fever.

B. Clinical Description

Rheumatic fever is a non-infectious, post-infection inflammatory disease affecting primarily the heart (causing valve damage), joints, subcutaneous tissues, and the central nervous system (CNS). It occurs as a delayed complication of infection with certain serotypes of GAS. Patients who have had rheumatic fever have a significantly elevated risk of recurrence of rheumatic fever following GAS infection, often with further cardiac damage. Another post-infection complication of GAS infection, usually following skin infection, is acute glomerulonephritis. This disease is associated with varying degrees of renal function impairment and protein in the urine.

C. Vectors and Reservoirs

Humans are the only known reservoir for *S. pyogenes*.

D. Modes of Transmission

The modes of transmission of GAS are large respiratory droplets and person-to-person transmission through direct contact with infected individuals or carriers. Indirect person-to-person spread through objects can sometimes occur. Nose, throat, skin, anal, and vaginal carriers can all serve as sources of GAS infection. Outbreaks of GAS pharyngitis may occur following ingestion of contaminated food (most commonly eggs, milk, and milk products).

E. Incubation Period

The incubation period for GAS infection is usually 1–5 days, rarely longer. Delayed sequelae constituting rheumatic fever occur, on average, 19 days after onset of preceding streptococcal pharyngitis (range of 1–5 weeks).

F. Period of Communicability or Infectious Period

In untreated, uncomplicated GAS infection, the infectious period starts several days before onset of symptoms and lasts from 10–21 days. If purulent discharge is present, the infectious period may be extended from weeks to months. Persons with untreated GAS pharyngitis may carry and transmit the bacteria for weeks or months, with sharply decreasing contagiousness 2–3 weeks after illness onset. The ability to transmit the organism is usually terminated within 24 hours of appropriate antibiotic treatment.

G. Epidemiology

Rheumatic fever occurs throughout the world and is seen more frequently in developing countries. There is seasonal variation in its occurrence, with more cases occurring during the late winter and early spring months, coincident with the occurrence of GAS pharyngitis. Cases of rheumatic fever are most commonly seen in children 3–15 years of age and rarely seen in infants and younger children. A person who has had rheumatic fever is at increased risk for recurrence of rheumatic fever after subsequent GAS infections.

H. Bioterrorist Potential

This pathogen is not considered to be of risk for use in bioterrorism.

Section 2:
REPORTING CRITERIA AND LABORATORY TESTING

A. What To Report to the Massachusetts Department of Public Health (MDPH)

Report any of the following:

- The onset of an inflammatory disease of the heart, joints, subcutaneous tissues, or CNS occurring 1–5 weeks after a GAS infection (with or without laboratory confirmation); or
- Inflammatory disease suggestive of rheumatic fever, without other likely explanation, associated with evidence of recent streptococcal infection on the basis of an antistreptolysin O (ASO) antibody or other anti-streptococcal antibodies.

Note: See Section 3C for information on how to report a case.

B. Laboratory Testing Services Available

The MDPH State Laboratory Institute (SLI), Reference Laboratory will test specimens for the presence of GAS when specimens are submitted as part of an epidemiologic investigation conducted by the MDPH. In some outbreak circumstances, isolates may be sent to the Centers for Disease Control and Prevention (CDC) for typing.

For more information on submitting specimens, contact the SLI Reference Laboratory at (617) 983-6607.

Section 3:
REPORTING RESPONSIBILITIES AND CASE INVESTIGATION

A. Purpose of Surveillance and Reporting

- To identify household contacts for culture and treatment.
- To identify infection sources of public health concern (e.g., contaminated food or a health care worker who is a GAS carrier), and to stop transmission from such sources.
- To monitor incidence of rheumatic fever to detect changes in characteristics of GAS infection.

B. Laboratory and Health Care Provider Reporting Requirements

Rheumatic fever is reportable to the local board of health (LBOH). The MDPH requests that health care providers immediately report to the LBOH in the community where the case is diagnosed, all confirmed or suspect cases of rheumatic fever, as defined by the reporting criteria in Section 2A.

Laboratories performing examinations on any specimens (from blood, CSF, or other normally-sterile body fluid only) derived from Massachusetts residents that yield evidence of GAS (*S. pyogenes*) infection shall report such evidence of infection directly to the MDPH within 24 hours.

C. Local Board of Health (LBOH) Reporting and Follow-Up Responsibilities

Reporting Requirements

MDPH regulations *(105 CMR 300.000)* stipulate that rheumatic fever is reportable to the LBOH and that each LBOH must report any confirmed case of rheumatic fever or suspect case of rheumatic fever, as defined by the reporting criteria in Section 2A. Cases should be reported to the MDPH Bureau of Communicable Disease Control, Office of Integrated Surveillance and Informatics Services (ISIS) using a MDPH *Generic Confidential Case Report Form* (found at the end of this chapter). Refer to the *Local Board of Health Timeline* at the end of this manual's *Introduction* section for information on prioritization and timeliness requirements of reporting and case investigation.

Case Investigation

1. It is the responsibility of the LBOH to complete a MDPH *Generic Confidential Case Report Form* (found at the end of this chapter) by interviewing the case and others who may be able to provide pertinent information. Much of the information required on the form can be obtained from the health care provider or from the medical record.
2. Use the following guidelines to assist in completing the form:
   a. Accurately record the case's demographic information.
   b. Accurately record clinical information including "rheumatic fever" as the disease being investigated, date of symptom onset, symptoms, whether hospitalized, and hospital and clinician contact information.
   c. Include all available diagnostic laboratory test information. Indicate the date of first positive GAS culture, if there was one. If other laboratory tests were used diagnostically (such as an ASO or other serologic test), please indicate the type(s) of test(s) used and the date(s) tested. Record this information in the "Comments" section at the bottom of the page, if necessary.
   d. Indicate the type of specimen from which GAS was isolated/identified (e.g., blood, cerebrospinal fluid [CSF]). Record this information in the "Comments" section at the bottom of the page.
   e. Include any additional comments regarding the case.
   f. If you have made several attempts to obtain case information but have been unsuccessful (e.g., the case or health care provider does not return your calls or respond to a letter, or the case refuses to divulge information or is too ill to be interviewed), please fill out the form with as much information as you have gathered. Please note on the form the reason(s) why it could not be filled out completely.

3. After completing the form, attach laboratory report(s) and fax or mail (in an envelope marked "Confidential") to ISIS. The confidential fax number is (617) 983-6813. Call ISIS at (617) 983-6801 to confirm receipt of your fax. The mailing address is:

MDPH, Office of Integrated Surveillance and Informatics Services (ISIS)
305 South Street, 5th Floor
Jamaica Plain, MA 02130
Fax: (617) 983-6813

4. Institution of disease control measures is an integral part of case investigation. It is the responsibility of the LBOH to understand, and if necessary, institute the control guidelines listed in Section 4.

Section 4:
CONTROLLING FURTHER SPREAD

A. Isolation and Quarantine Requirements *(105 CMR 300.200)*

Minimum Period of Isolation of Patient

Until 24 hours after initiation of antibiotic therapy.

Minimum Period of Quarantine of Contacts

Search for carriers among close contacts and treat them with antibiotics to prevent other cases of GAS infection and rheumatic fever. Otherwise, no restrictions.

B. Protection of Contacts of a Case

Siblings and other household contacts of a case should have throat cultures taken, and if positive for GAS, be treated with antibiotics. Other close contacts (those in contact with the case's respiratory secretions) should be evaluated and cultured, as indicated.

C. Managing Special Situations

Daycare

Consider throat cultures for all symptomatic daycare attendees and staff who are close contacts (i.e., those in contact with the case's respiratory secretions), with subsequent antibiotic treatment of those found to be GAS-culture positive. Contact the MDPH Division of Epidemiology and Immunization, at (617) 983-6800 or (888) 658-2850, for assistance in managing follow-up of a case of rheumatic fever in a daycare setting.

School

Consider throat cultures for all symptomatic classroom members and other close contacts (i.e., those in contact with the case's respiratory secretions), with subsequent antibiotic treatment of those found to be GAS-culture positive. Contact the MDPH Division of Epidemiology and Immunization, at (617) 983-6800 or (888) 658-2850, for assistance in managing follow-up of a case of rheumatic fever in a school setting.

Reported Incidence Is Higher Than Usual/Outbreak Suspected

If the number of reported cases of rheumatic fever or serious GAS infections in your city/town is higher than usual or if you suspect an outbreak, contact the epidemiologist on-call at the MDPH Division of Epidemiology and Immunization at (617) 983-6800 or (888) 658-2850. The Division can help determine a course of action to prevent further cases and can perform surveillance for cases across town lines, which would otherwise be difficult to identify at the local level.

D. Preventive Measures

Environmental Measures

Advise daycare centers to clean toys daily using an EPA-registered disinfectant, and to discourage the use of play food, which facilitates the transmission of this and many other agents.

Personal Preventive Measures/Education

Advise individuals to:

- Practice good personal hygiene, emphasizing the need for proper handwashing.
- Avoid sharing food, drinks, cigarettes, eating or drinking utensils, or other personal items such as lip gloss.

A Group A Streptococcal Disease Public Health Fact Sheet is available from the MDPH Division of Epidemiology and Immunization or on the MDPH website at www.mass.gov/dph. Click on the "Publications and Statistics" link, and select the "Public Health Fact Sheets" section under "Communicable Disease Control."

ADDITIONAL INFORMATION

The following is the formal CDC surveillance case definition for rheumatic fever. It is provided for your information only and should not affect the investigation and reporting of a case that fulfills the criteria in Section 2A of this chapter. (The CDC and the MDPH use the CDC case definitions to maintain uniform standards for national reporting.) For reporting to the MDPH, always use the criteria outlined in Section 2A.

Note: The most up-to-date CDC case definitions are available on the CDC website at www.cdc.gov/epo/dphsi/casedef/case_definitions.htm.

Clinical Description

An inflammatory illness that occurs as a delayed sequela of GAS infection.

Major Criteria

Carditis, polyarthritis, chorea, subcutaneous nodules, and erythema marginatum.

Minor Criteria

- Previous rheumatic fever or rheumatic heart disease;
- Arthralgia;
- Fever;
- Elevated erythrocyte sedimentation rate, positive C-reactive protein, or leukocytosis; and
- Prolonged PR interval on an electrocardiogram.

Laboratory Criteria for Diagnosis

No specific laboratory test exists for the diagnosis of rheumatic fever.

Confirmed	An illness characterized by: a) two major criteria or one major and two minor criteria (as described in *Clinical Description* section); and b) supporting evidence of preceding GAS infection.

Comment

Supporting evidence to confirm streptococcal infection includes increased ASO or other streptococcal antibodies, throat culture positive for GAS, or recent scarlet fever. The absence of supporting evidence of preceding streptococcal infection should make the diagnosis doubtful, except in Sydenham chorea or low-grade carditis, when rheumatic fever is first discovered after a long latent period from the antecedent infection.

REFERENCES

American Academy of Pediatrics. [Group A Streptococcal Infections.] In: Pickering L.K., ed. *Red Book: 2003 Report of the Committee on Infectious Diseases, 26th Edition.* Elk Grove Village, IL, American Academy of Pediatrics; 2003: 581–583.

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Heymann, D., ed. *Control of Communicable Diseases Manual, 18th Edition*. Washington, DC, American Public Health Association, 2004.

Mandell, G., Bennett, J., Dolin, R., eds. *Principles and Practice of Infectious Diseases, 6th Edition.* New York, Churchill Livingstone Inc., 2004.

MDPH. *Regulation 105 CMR 300.000: Reportable Diseases, Surveillance, and Isolation and Quarantine Requirements*. MDPH, Promulgated November 4, 2005.

FORMS & WORKSHEETS

Rheumatic Fever

Rheumatic Fever

LBOH Action Steps

This form does not need to be submitted to the MDPH with the case report form. It is for LBOH use and is meant as a quick-reference guide to rheumatic fever case investigation activities.

LBOH staff should follow these steps when rheumatic fever is suspected or confirmed in the community. For more detailed information, including disease epidemiology, reporting, case investigation, and follow-up, refer to the preceding chapter.

- ❑ Notify the MDPH Division of Epidemiology and Immunization, at (617) 983-6800 or (888) 658-2850, to report any confirmed or suspect case(s) of rheumatic fever.
- ❑ Obtain laboratory confirmation.
- ❑ Determine if the case attends daycare or school, and if so, contact the MDPH Division of Epidemiology and Immunization for case management assistance.
- ❑ Identify other potentially-exposed persons.
- ❑ Fill out the case report form (attach laboratory results).
- ❑ Send the completed case report form (with laboratory results) to the MDPH Bureau of Communicable Disease Control, Office of Integrated Surveillance and Informatics Services (ISIS).